Neuroendocrine Tumors

Neuroendocrine tumors affect cells throughout the nervous and endocrine systems that produce and secrete regulatory hormones. Common sites of origin include the endocrine pancreas; parathyroid, adrenal, and pituitary glands; calcitonin-producing cells of the thyroid (causing medullary thyroid carcinoma); and argentaffin cells of the gut (causing carcinoid tumors). Neuroendocrine tumors are rare and can be broadly subdivided into those with and those without a clinical syndrome. Most neuroendocrine tumors are malignant and metastasize commonly to lymph nodes and the liver or less commonly to bone, lung, brain, and other organs. Despite the widespread metastasis, these tumors are typically slow-growing and often have an insidious presentation. These guidelines discuss the diagnosis and management of neuroendocrine tumors and special considerations relating to these tumors; important updates include expanded information on octreotide use and a new algorithm on neuroendocrine tumors of unknown primary.